**PATENT**
Attorney Docket No. 24414-Y

**IN THE UNITED STATES PATENT AND TRADEMARK OFFICE**

In re Application of:

Arnold J. REUSER et al.

Serial No: n/a

Filed: Dec. 14, 2001

For: **COMPOSITIONS AND METHODS FOR TREATING ENZYME DEFICIENCY**

**PRELIMINARY AMENDMENT**

Commissioner for Patents
Washington, D.C. 20231

Sir:

Before action in this application, please amend the above-identified application as follows:

**IN THE CLAIMS**

Please amend claims 29-34 and 52-53 and add new claims 62-67 as indicated in Appendices A and B submitted herewith. Appendix A is a marked-up copy of the amended claims and Appendix B is a clean copy of the amended claims.

**REMARKS**

Claims 1-67 are currently pending in the present application. The amendments do not add any new matter under 35 U.S.C. §132. The claims have been amended to remove multiple dependencies. Basis for the term "recombinant human acid alpha

glucosidase containing mannose 6-phosphate" can be found on page 23, lines 5-6 and 10 of the original specification as filed. Basis for the term "present at a level of at least 50 ug/ml" can be found on page 30, line 22 of the original specification as filed. Basis for the term "mannose 6-phosphare containing lysosomal protein" can be found on page 1, lines 13-14 of the original specification as filed. Basis for the term "phosphorylated at the 6' position of its mannose group" can be found on page 9, lines 24-25 of the original specification as filed. Accordingly, entry of the amendments prior to examination of the application is respectfully requested.

Respectfully submitted,

**NATH & ASSOCIATES PLLC**

Date: Dec 14, 2001

**NATH & ASSOCIATES PLLC**
1030 Fifteenth Street, N.W.
Sixth Floor
Washington, D.C. 20005-1503
Telephone: (202) 775-8383
Facsimile: (202) 775-8396
TLJ:JBG:\prelim.doc

Todd L. Juneau
Reg. No. 40,669
Customer No. 20529

**BOX PATENT**
Attorney Docket No. 24414-Y

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## Appendix A

Please amend the following claims as indicated in the following marked up copy of the claims.

29. (Once Amended) The method of [any of claims 21-28] claim 28, wherein the patient is administered a single dosage of alpha-glucosidase per week.

30. (Once Amended) The method of [any of claims 21-28] claim 21, wherein the patient is administered two dosages of alpha-glucosidase per week.

31. (Once Amended) The method of [any of claims 21-28] claim 21, wherein the patient is administered three dosages of alpha-glucosidase per week.

32. (Once Amended) The method of [any of claims 21-31] claim 21, wherein the amount is administered per week for a period of at least four weeks.

33. (Once Amended) The method of [any of claims 21-31] claim 21, wherein the amount is administered per week for a

period of at least 24 weeks.

34. (Once Amended) The method of [any of claims 21-31] claim 21, wherein the alpha-glucosidase was produced in milk of a transgenic animal.

52. (Once Amended) The method of [any one of claims 47-51] claim 47, wherein the first, second, third and fourth dosages are each administered for periods of 15 min to 8 hours.

53. (Once Amended) The method of [any one of claims 47-51] claim 47, wherein the first, second, third and fourth dosages are administered for periods of 1 hr, 1hr, 0.5 hr and 3 hr respectively.

Please add the following new claims.

62. A pharmaceutical composition comprising recombinant human acid alpha glucosidase containing mannose 6-phosphate and a pharmaceutically acceptable carrier.

63. The pharmaceutical composition of claim 62, wherein the recombinant human acid alpha glucosidase containing mannose 6-phosphate is present at a level of at least 50 µg/ml.

64. A pharmaceutical composition comprising a purified mannose 6-phosphate containing lysosomal protein and a pharmaceutically acceptable carrier, wherein the lysosomal protein is recombinant human acid alpha glucosidase.

65. The pharmaceutical composition of claim 64, wherein the mannose 6-phosphate containing lysosomal protein is present at a level of at least 50 µg/ml.

66. A pharmaceutical composition comprising recombinant human acid alpha glucosidase phosphorylated at the 6′ position of its mannose group and a pharmaceutically acceptable carrier.

67. The pharmaceutical composition of claim 66, wherein said recombinant human acid alpha glucosidase phosphorylated at the 6′ position of its mannose group is present at a level of at least 50 µg/ml.

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For: **COMPOSITIONS AND METHODS FOR TREATING ENZYME DEFICIENCY**

**Appendix B**

Please amend the following claims as indicated in the following marked up copy of the claims.

29. (Once Amended) The method of claim 28, wherein the patient is administered a single dosage of alpha-glucosidase per week.

30. (Once Amended) The method of claim 21, wherein the patient is administered two dosages of alpha-glucosidase per week.

31. (Once Amended) The method of claim 21, wherein the patient is administered three dosages of alpha-glucosidase per week.

32. (Once Amended) The method of claim 21, wherein the amount is administered per week for a period of at least four weeks.

33. (Once Amended) The method of claim 21, wherein the amount is administered per week for a period of at least 24

weeks.

34. (Once Amended) The method of claim 21, wherein the alpha-glucosidase was produced in milk of a transgenic animal.

52. (Once Amended) The method of claim 47, wherein the first, second, third and fourth dosages are each administered for periods of 15 min to 8 hours.

53. (Once Amended) The method of claim 47, wherein the first, second, third and fourth dosages are administered for periods of 1 hr, 1hr, 0.5 hr and 3 hr respectively.

Please add the following new claims.

62. A pharmaceutical composition comprising recombinant human acid alpha glucosidase containing mannose 6-phosphate and a pharmaceutically acceptable carrier.

63. The pharmaceutical composition of claim 62, wherein the recombinant human acid alpha glucosidase containing mannose 6-phosphate is present at a level of at least 50 µg/ml.

64. A pharmaceutical composition comprising a purified mannose 6-phosphate containing lysosomal protein and a pharmaceutically acceptable carrier, wherein the lysosomal protein is recombinant human acid alpha glucosidase.

65. The pharmaceutical composition of claim 64, wherein the mannose 6-phosphate containing

lysosomal protein is present at a level of at least 50 µg/ml.

66. A pharmaceutical composition comprising recombinant human acid alpha glucosidase phosphorylated at the 6′ position of its mannose group and a pharmaceutically acceptable carrier.

67. The pharmaceutical composition of claim 66, wherein said recombinant human acid alpha glucosidase phosphorylated at the 6′ position of its mannose group is present at a level of at least 50 µg/ml.